Pediatric Palliative Care: Current Evidence and Evidence Gaps

Elissa G. Miller, MD1,2, Carly Levy, MD1,2, Jennifer S. Linebarger, MD3, Jeffrey C. Klick, MD4, and Brian S. Carter, MD3

Hospice and palliative medicine (HPM) is an emerging medical subspecialty initially recognized by the American Board of Medical Specialties in 2006. As the Institute of Medicine reported in 2014, health care delivery and the challenges of care at the end of children’s lives have evolved significantly over the past 15 years. The last decade has seen the growth of pediatric palliative care (PPC) programs at children’s hospitals and a transformation of the field. Twenty percent of children’s hospitals nationwide have a PPC program, with a peak in new program development occurring in 2008. New programs are almost immediately impactful, experience high referral volumes, and must quickly expand their workforce. Although HPM has a growing body of literature, the evidence for PPC appears to lag behind the clinical growth.

Here we provide a focused and practical summary of evidence in pediatric HPM as a primer for those practicing in other subspecialties. Specifically, we review evidence by focusing on a few of the domains in HPM: (1) communication and psychosocial support; (2) pain and symptom management; (3) end-of-life care; and (4) ways to build a better health care system. We hope to strengthen understanding and partnerships between HPM and non-HPM clinicians and investigators and thereby improve patient care for children with serious illnesses.

Case 1

An 18-day-old, term infant presented to the emergency department unresponsive with sepsis and presumed meningitis. Blood and cerebrospinal fluid cultures confirmed group-B streptococcus infection. Given the serious nature of the infant’s condition, the pediatric intensive care unit (PICU) team consulted the PPC team on hospital day 3. With aggressive life-sustaining measures, the infant survived, but sustained significant brain injury, remaining comatose with absent gag reflex and insufficient respiratory effort.

The PPC team met with family frequently during the infant’s hospitalization to provide support around understanding of disease and coping with critical illness as well as to address goals of care. Initially, goals were focused on life-sustaining measures. As the severity of the infant’s neurologic condition became clear, family goals slowly shifted toward a comfort-focused approach. Ultimately, after a family meeting with extended family present, the parents chose to focus solely on the infant’s comfort and discontinue all interventions that were prolonging suffering, including the ventilator. The infant died peacefully in his mother’s arms shortly thereafter.

Case 2

A 15-year-old male with Hunter syndrome was well known to his hospital’s PPC team. Many years earlier, his family, knowing the progressive nature of the disease, had chosen to focus on his comfort and quality of life. Enzyme infusions successfully reduced his pain and symptom burden, and he was rarely hospitalized during his first decade of life. Recently, however, he had experienced increased seizures with frequent aspiration events. After an episode of severe pneumonia requiring hospitalization and noninvasive ventilation, his parents asked to speak with the PPC team. Together, they devised an advance care plan for their son that included an out-of-hospital do not resuscitate (DNR) order. Utilizing the Concurrent Care for Children Requirement from the Affordable Care Act, the family chose to continue enzyme infusions while he also received home hospice care. Nine months later, the child had not been rehospitalized and was doing well, so the family discontinued hospice care but maintained the out-of-hospital DNR order. His parents continue to focus on his comfort and quality of life.

These cases present different but common scenarios facing PPC teams. A multidisciplinary approach to care allowed 2 different children—one child with sudden, critical illness and another with chronic disease—to live what their families felt was the best quality of life for as long as possible under the circumstances of their illness. It is important to examine the evidence guiding the practices discussed in these cases.

Communication and Psychosocial Support

The literature within the communication domain of HPM focuses on family meetings, decision making, advance care planning (ACP), family coping, and the impact of bereavement. Although there is significant literature focused on adult patients, pediatrics necessitates a stronger focus on the child’s cognitive development and how he or she fits into...
the social structure of the family. These differences make it difficult to extrapolate from the adult-based literature.

Family Meetings
As in our case 1 example, family meetings are a cornerstone tool of HPM, but they have been minimally studied in pediatrics. Nonetheless, it seems clear that these meetings are infrequently used, even for the sickest children. In one study, meetings were focused on information exchange and future management. Conferences occurred predominantly for sicker patients, those with complex chronic conditions, and those with PPC consultation. Although many clinicians believe that the family meeting is key to the practice of palliative medicine, there is minimal evidence regarding families’ perception of such meetings.

Decision Making
Clinicians can facilitate families’ decision making. Hinds et al interviewed parents faced with making noncurative treatment decisions. The families identified components of being a “good parent” to include focusing on their child’s quality of life, advocating for their child with the medical team, and putting their child’s needs above their own. By working to define a family’s priorities and values, the clinician can help them fulfill their wishes to “be a good parent” through dire circumstances. In a study of parents of PICU patients, 40% of parents preferred sharing decision making with their doctors, 41% preferred autonomous decision making, and 18.9% preferred delegating these decisions to their doctors.

ACP
ACP involves the patient, family, and providers sharing information on the natural course of the child’s disease, individualized prognostic information including trajectory, and the expected experiences of the child. ACP is a common task for palliative care teams; however, conversations are most productive when the family and clinicians have a common understanding of the child’s disease and prognosis. On average, oncologists recognize 100 days sooner than parents that there is no realistic chance of cure. When the poor prognosis for survival is disclosed early, there are earlier discussions of hospice care, earlier DNR orders, and decreased use of chemotherapy in the last month of life. Importantly, there are also better parental ratings of the quality of care in the home.

It is also important to include the child in ACP in appropriate ways. Teenagers with HIV/AIDS and those with advanced cancer overwhelmingly express the desire to participate in their own ACP. Families who decide to speak with their children about death report that they are happy with that decision, and nearly one-third of families who did not discuss death with their child regretted that decision later.

A single-center study examined generally perceived barriers to ACP and DNR discussions. The top 3 barriers to ACP discussions identified were clinician perception of unrealistic parental expectations, lack of parent readiness to have the discussion, and discrepancies between clinician and patient/parent understanding of the prognosis. The clinicians at that center overwhelmingly felt that ACP/DNR discussions should be initiated on presentation/diagnosis or during a period of stability.

Family Coping
Families cope and accommodate to their child’s illness using a multitude of supports, coping strategies, and resources. Religion, spirituality, and life philosophy are some tools that can help families cope with adversity. Hopeful thoughts and language play a major role; many parents use hopeful language about outcomes for their child, even when death is inevitable.

Impact of Bereavement
The death of a child or sibling has a significant effect on the entire family. Youngblut et al reported significant negative physical and mental health outcomes in parents 13 months after their child’s death, including newly diagnosed chronic health conditions, posttraumatic stress disorder, and hospitalizations. Psychiatric comorbidities, previous loss, economic hardship, duration and intensity of the child’s treatment, perceptions of medical care, child’s quality of life, and parent preparedness for death all impact parental outcomes. Siblings are affected as well and often recognize their parents’ grief. Survivor guilt, parental overprotection, and idealization of the deceased child affect sibling bereavement. Siblings may be affected by how their parents engage them, allow them to see/hold/touch their ill siblings, and even participate in their care.

Physical Aspects of Care
In contrast to the large body of evidence on palliative pain and symptom management in adults, studies on pediatric patients are scant. Much of the evidence that pediatric HPM clinicians use is drawn from the adult literature, and the medications used are often off-label. Although many of the principles of pain management are consistent across the age spectrum, there are important differences in neonatal and pediatric patients that merit more careful study. Even though the clinical practice of PPC includes symptom management integrated into life-sustaining therapies, much of the PPC literature focuses on pain and symptom management at the end of life.

Symptoms at End of Life
Children with terminal cancer have a significant burden of pain and symptoms in their last 30 days of life. PPC has been shown to lessen this burden, and to help parents feel more prepared during their child’s last months of life and at the time of death. Prominent symptoms in the last 24 hours of life include respiratory distress, pain, nausea, vomiting, and anxiety. One study found that children who die of advanced
heart disease also carry a significant symptom burden and poor quality of life in their last 30 days, suggesting the need for improved symptom assessment and management in many children with noncancer diagnoses.

**End-of-Life Care**

**Resuscitation Status**
In a study of deaths of hospitalized children at 1 institution, few children had DNR orders before admission, and often orders that were written were done so less than 1 day before death. The incidence of DNR orders increased when parents had more education (>15 years) or a higher monthly income. On the other hand, a recent report on PICU deaths suggested that DNR orders may be used less often today than in the past. We do not know whether PPC involvement increases the frequency of DNR or limited code orders.

**How and Where Patients Die**
As access to PPC has increased, the location of death for children dying from oncologic diseases and complex chronic conditions has shifted. Nonetheless, children with advanced heart, lung, immune, and infectious diseases continue to die in the PICU after receiving highly technical medical support at the end of life and after the withdrawal of life-sustaining interventions. Patients and families vary in terms of preference for location of end-of-life care, and the majority of children die in a hospital setting. This can actively change to home deaths with PPC involvement. Among patients with advanced cancer, having open discussions about the location of death is associated with increased home deaths. Although adult HPM providers use a shift in location of death as an impactful outcome of palliative care involvement, whether PPC will, or even should, have the same impact is less clear. Many children and families become comfortable with the hospital environment over the disease trajectory and actively choose the hospital for end-of-life care.

**Building a Better Health Care System**

**Barriers to Palliative Care**
A survey of pediatric staff members at the University of California San Francisco identified an uncertain patient prognosis as the primary barrier to PPC referral. Additional barriers included the clinician’s perception of the family’s unwillingness to acknowledge the incurable nature of their child’s disease, language barriers, and time constraints. Many pediatricians still associate PPC with hospice care, used only for the end of life. In reality, most PPC consults are for facilitating communication; in 1 study, only 30% of consults ended in the patient’s death.

**Cost of Care**
By clarifying and focusing on individual patient goals of care, adult palliative care teams not only save money by decreasing hospital length of stay, but also help reduce 30-day hospital readmission rates and decrease emergency department and intensive care unit utilization. Pediatric data are currently lacking, however, and some argue that cost avoidance is not a relevant topic in PPC.

Children with life-limiting diseases experience multiple hospitalizations and spend on average more than 39 days in the hospital in the last year of life. Outpatient PPC programs can decrease hospital and emergency department charges and shift care to an in-home setting through a community-based PPC program. However, to date no study has examined the overall health care costs and potential savings realized by PPC involvement with medically complex hospitalized pediatric patients.

**Care Delivery Systems**
There are multiple reports of novel models of PPC delivery, from perinatal palliative care to automated PPC consult triggers to the use of teledicine. Pediatric hospice patients are eligible for concurrent care (ie, hospice in addition to disease-directed therapy, as in our case 2) through the Affordable Care Act. The use of concurrent care has been reported, but cost savings remain unclear.

**Use of the PPC Team**
Although palliative care offers a variety of expertise, previous studies have demonstrated that the specific services requested of the palliative care team vary and depend largely on the referring service. A survey of PICU physicians identified continuity between hospitalizations, transition to the community setting, and provision of bereavement care for family and staff was the most valuable services offered by the PPC team. Critical care physicians were more likely to consult palliative care for psychosocial support of patients and families, and oncologists were more likely to consult for symptom management. In addition, clinicians with self-reported palliative care education were more likely to involve PPC in the care of their patients.

**Best Practices/Quality Improvement Measures**
Most notably lacking from the PPC literature are data confirming quality metrics and establishing evidence-based best practices for PPC clinicians.

**Barriers to PPC Research**
There are many barriers to producing high-quality palliative care research, including funding, institutional capacity, researcher workforce, challenges related to the topic and population (eg, attrition, rare disease entities, heightened human subjects protections), and public and professional misunderstanding of palliative care and aversion to topics related to serious illness and the end of life. It is important to focus PPC research on areas that are unique to pediatric patients (eg, developmentally appropriate communication, family coping), those that are unique to PPC (eg, concurrent hospice care, care coordination), and those that cannot be easily extrapolated from adult literature (eg, pain and symptom management).
Discussion

The evidence shows that PPC teams improve patient care, and it is no longer acceptable for pediatric patients with access to PPC to die without PPC involvement. It is striking that data on costs of care, best practices, and management lag behind those for ACP.

Ulrich and Morrison suggested opportunities for collaboration for future research, stating that PPC “research priorities...should fall into one of two categories. They should be either issues related specifically to children, from which lessons learned from adult research cannot be extrapolated, or unique opportunities for PPC research to inform the provision of palliative care for people of all ages.” The Institute of Medicine’s 2014 report echoes this sentiment. Pediatric-specific research efforts intended to directly impact clinical care should focus on such areas as developmental considerations to optimize patient and family communication, evidence-based interventions to maximally support family members and aid in decision making, families’ perspective on family meetings, and models for delivery of out-of-hospital PPC (Table; available at www.jpeds.com).

Broad application of the current evidence should result in decreased patient suffering and less discrepancy in palliative care between adult and pediatric patients. Such application across pediatric subspecialties will allow clinical and research collaboration to fill in the disease-specific evidence gaps that remain. With approximately 250 pediatricians board-certified in HPM, we have few practitioners of clinical PPC and even fewer clinician-investigators. New HPM fellows are graduating from 1-year clinical HPM training programs and generally are not being trained to become academic clinician-investigators; consequently, growth of the evidence base for PPC likely will continue to rely on relatively few pediatric investigators and to lean heavily on other fields as partners in advancing this young field.

Submitted for publication Nov 12, 2014; last revision received Jan 30, 2015; accepted Feb 6, 2015.

Reprint requests: Elissa G. Miller, MD, Nemours/Alfred I. duPont Hospital for Children, PO Box 269, Wilmington, DE 19899. E-mail: elissa.miller@nemours.org

References


GRAND ROUNDS

June 2015

Pediatric Palliative Care: Current Evidence and Evidence Gaps

1539
<table>
<thead>
<tr>
<th>Topic</th>
<th>Research progress to date</th>
<th>Proposed needed investigation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Communication and psychosocial support</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Family meetings</td>
<td>• Family meetings rarely occur.</td>
<td>• Ideal timing/frequency of family meetings</td>
</tr>
<tr>
<td></td>
<td>• Effective in aiding decision making.</td>
<td>• Whether routine family meetings affect patient-level outcomes (eg, length of stay, family and/or provider satisfaction with care)</td>
</tr>
<tr>
<td></td>
<td>• An evidence-based structure for family conferences exists.</td>
<td>• Evidence-based methods of providing decision support, including how to clearly communicate patient’s clinical status, review the benefits/harms of medical interventions, and discuss goals of care</td>
</tr>
<tr>
<td>Decision making</td>
<td>• Families differ in their approach to decision making (shared vs autonomous vs delegation).</td>
<td></td>
</tr>
<tr>
<td></td>
<td>• Parents differ in what it means to be a “good parent,” making it challenging to best to support families with difficult treatment decisions.</td>
<td></td>
</tr>
<tr>
<td>ACP</td>
<td>• Teens want to participate in ACP.</td>
<td>• Best time for prognostic revelation and ACP discussions</td>
</tr>
<tr>
<td></td>
<td>• Teens feel marginalized when excluded from ACP discussions.</td>
<td>• Best ways to communicate and document ACP discussions</td>
</tr>
<tr>
<td>Family coping</td>
<td>• Families rely on various coping strategies when their child has a serious illness.</td>
<td>• Evidence-based strategies for providers to elicit and support family coping strategies</td>
</tr>
<tr>
<td>Bereavement support</td>
<td>• Families use spirituality in their coping after the death of a child, although they often have negative physical and mental health outcomes.</td>
<td>• Complicated bereavement risk-group identification</td>
</tr>
<tr>
<td></td>
<td>• Siblings are affected as well.</td>
<td>• Evidence-based parent and sibling bereavement interventions</td>
</tr>
<tr>
<td></td>
<td>• There are few evidence-based bereavement interventions for siblings.</td>
<td></td>
</tr>
<tr>
<td>Physical aspects of care</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pain and symptom management</td>
<td>• The most distressing symptoms have been identified by patients and families.</td>
<td>• Pharmacologic and nonpharmacologic interventions merit robust study, including effectiveness of different medications and potential problems of polypharmacy</td>
</tr>
<tr>
<td></td>
<td>• All palliative symptom management research to date has been in adult populations.</td>
<td></td>
</tr>
<tr>
<td>Symptoms at end of life</td>
<td>• PPC helps decrease patients’ suffering at end of life and improves parents’ sense of preparedness before the death of the child.</td>
<td>• Tools for symptom assessment in non–cancer patient populations</td>
</tr>
<tr>
<td>End of life care</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Resuscitation status</td>
<td>• DNR orders are commonly placed just prior to a child’s death.</td>
<td>• Whether the involvement of a PPC team leads to earlier limits on resuscitative efforts</td>
</tr>
<tr>
<td></td>
<td>• Death in the PICU continues to be common in children after withdrawal of technological support.</td>
<td>• Technology-based decision aids (eg, videos, apps) for parents and pediatric patients</td>
</tr>
<tr>
<td>How and where patients die</td>
<td>• PPC teams help shift the location of death away from PICU and inpatient settings to home and hospice settings.</td>
<td>• Identification and clarification of factors that influence the preferred location of death</td>
</tr>
<tr>
<td>Building a better healthcare system</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Barriers to palliative care</td>
<td>• Uncertain patient prognosis.</td>
<td>• Comparison of clinician perception of parent readiness to engage with PPC vs parent report of readiness</td>
</tr>
<tr>
<td></td>
<td>• Clinician perception of family “readiness” for PPC/ACP discussions.</td>
<td>• Interventions to facilitate earlier PPC involvement and earlier ACP</td>
</tr>
<tr>
<td></td>
<td>• Misperception that palliative care is equivalent to hospice or only for the end of life.</td>
<td></td>
</tr>
<tr>
<td>Cost of care</td>
<td>• Adult palliative care programs reduce emergency department and intensive care unit utilization, decrease hospital length of stay, and cut costs, as do outpatient PPC programs.</td>
<td>• Overall health care cost and potential savings of PPC involvement with medically complex, hospitalized pediatric patients</td>
</tr>
<tr>
<td>Care delivery systems</td>
<td>• There are multiple proof-of-concept descriptions, but no studies of novel modes of PPC delivery.</td>
<td>• Cost of PPC team (eg, salaries) weighed against healthcare dollars saved through outpatient PPC</td>
</tr>
<tr>
<td>Use of the PPC team</td>
<td>• Clinicians from different subspecialties consult PPC for different reasons.</td>
<td>• Evidence-based PPC team structure/staffing</td>
</tr>
<tr>
<td></td>
<td>• Clinicians with self-reported PPC education are more likely to involve PPC in the care of their patients.</td>
<td>• Effect of concurrent care on cost of care, family perceptions of care, and treatment decisions</td>
</tr>
<tr>
<td>Best practices/quality improvement measures</td>
<td>• None exist.</td>
<td>• Evidence-based educational models for practicing physicians to improve understanding and use of PPC</td>
</tr>
<tr>
<td>Barriers to PPC research</td>
<td>• Barriers to PPC research include lack of funding, institutional capacity, and research workforce.</td>
<td>• Evidence-based quality metrics</td>
</tr>
<tr>
<td></td>
<td>• Additional challenges include public and professional misunderstanding of the field.</td>
<td>• Evidence-based best practice guidelines</td>
</tr>
</tbody>
</table>